– increased plasma levels of BNP in 45.7% of patients, to value 240 ng/ml increasing cTnI values plasma at 4.34% of cases.

Biological changes were correlated in most cases with clinical manifestations, echo changes induced cardiotoxicity and increase of dispersion QT/QTc intervals.

Conclusions Increased levels of cardiac biomarkers: BNP and cTnI and of the dispersion of QT/QTc intervals in children treated with anthracyclines±other drugs with cardiotoxic efects positively correlates with installation of the cardiotoxicity with clinical or infraclinical manifestations, constituing an useful indicator for the.cardiotoxicity. Changes in this parameters appeared early than echo changes anthracicline induced cardiotoxicity and is necessary to systematic monitoring these parameters during and after cytostatic treatment.

The author hereby declares no conflict of interest

0454

Long-term cardiac prognosis and risk stratification in 260 adults presenting with mitochondrial diseases

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Aims To assess the long-term cardiac prognosis of adults with mitochondrial diseases.

Methods and results Between January 2000 and May 2014, we retrospectively included in this study 260 consecutive patients (60% women) ≥18 years of age, (interquartile range [IQR]; 31 to 54), with genetically proven mitochondrial diseases, including 109 with mitochondrial DNA (mtDNA) single large-scale deletions, 64 with the m.3243A>G mutation in MT-TL1, 51 with other mtDNA point mutations, and 36 patients with nuclear genes mutations. Cardiac involvement was present at baseline in 81 patients (30%). Single and multiple variable analyses were performed in search of predictors of major adverse cardiac event (MACE), and hazard ratios (HR) and 95% confidence intervals (CI) were calculated. Over a median follow-up of 7 years [3.6 to 11.7], 27 patients (10%) suffered a MACE, defined as sudden death, death due to heart failure (HF), resuscitated cardiac arrest, 3rd degree atrioventricular block, sinus node dysfunction, cardiac transplantation, or hospitalization for management of HF. Patients with single large-scale mtDNA deletions or m.3243A>G mutations had the highest incidence of MACE. By multiple variable analysis, intraventricular conduction block (HR=16.9; 95% CI: 7.2 to 39.4), diabetes (HR=7.0; 95% CI: 2.9 to 16.7), premature ventricular complexes (HR=3.6; 95% CI: 1.4 to 9.2) and left ventricular (LV) hypertrophy (HR=2.5; 95% CI: 1.1 to 5.8) were independent predictors of MACE. In patients with 0, 1, and ≥2 risk factors, the incidence of MACE was 1.7, 15 and 42% respectively.

Conclusions Patients with mitochondrial diseases are at high risk of MACE, independently predicted by intraventricular conduction block, diabetes, ventricular prematurity and LV hypertrophy.

The author hereby declares no conflict of interest

0353

Prevalence of hereditary transthyretin cardiac amyloidosis in patients with increase in LV thickness in France

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Background Hereditary transthyretin cardiac amyloidosis (mTTR-CA) is a hypertrophic cardiomyopathy with challenging diagnosis and poor prognosis. The prevalence of m-TTR in patients with increased left ventricular wall thickness (LVWT) is unknown.

Methods Prospective and consecutive multicenter study with systematic genetic screening for mTTR in adult patients with LVWT ≥15mm included in cardiology primary clinics.

Results 298 patients were genotyped of whom 23% were African descendant. The median (IQR) age was 62(50,74), 74% were men and 36% were in NYHA class III-IV. The median of maximal LV thickness was 18 (16, 21)mm.17 patients had TTR mutation (5.7%) of whom 15 (5.0) had confirmed mTTR-CA. All the mTTR-CA were ≥55years meaning that the prevalence of mTTR-CA was 8.3% above this age. Of the 15 with mTTR-CA, 8 were Africans and 6 Caucasians. In Africans ≥55 years, the prevalence was 22% and reached 35% in those over 65 years. The most frequent mutations were V142I (8), V50M (2) and I127V (2).

When adjusted to age, neuropathy (OR=20.1; 95%-CI, 5.86-69.4; P<0.001), carpal tunnel syndrome (OR=15.31; 95%-CI, 4.32-54.3; P<0.001), ECG low voltage (OR=8.8; 95%-CI, 2.67-29.1; P<0.001), symmetric hypertrophy (OR=10.9; 95%-CI, 1.97-59.8; P=0.006), LVEF impairment (OR=10.9; 95%-CI, 1.62-15.5; P=0.005), and late gadolinium enhancement at MRI (OR=42.9; 95%-CI, 2.38-772; P=0.011) were all associated with increased odds of CA.

Conclusions mTTR-CA is frequent in HCM, particularly in African descendant and patients ≥55 years. mTTR genetic screening may be warranted for patients with increased LVWT, especially with neuropathy or carpal tunnel syndrome or LGE at MRI.

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0161

Usefulness of plasma high sensitive troponin t and Nt-proBNP in the diagnosis of cardiopathy in Friedreich ataxia

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Friedreich ataxia, due to mitochondrial dysfunction, is the most common genetic sensory ataxia. It's due to lack of frataxin. Hypertrophic cardiomyopathy is associated with Friedreich ataxia and is the major cause of death, (80%).

This study concerned the role of plasma biomarkers, high sensitive troponin and Nt-proBNP, in the diagnostic of cardiopathy in Friedreich ataxia.

From December 2012 to January 2015, we included 76 genetically confirmed Friedreich's ataxia patients in Pitié-Salpêtrière Hospital. Clinical examination, ECG, echocardiography and blood samples were obtained.

Patients were aged of 38±12 years, (mean±sd), 50% were male. 4 patients had palpitations, 2 dyspnea and no patients had chest pain. 89% had negative T waves on the ECG. 49% had echographic cardiac hypertrophy according to Henry's nomogram. Patients with hypertrophy were younger: 34±10 years versus 42±14 years, age at onset of the disease was earlier: 15±6 years versus 21±15 years. Interventricular septal wall thickness was 12,9±1,7mm versus 10±1,2mm, and posterior wall thickness was 11,3±1,5mm versus 9,4±1mm. Left ventricle ejection fraction was similar (64%). For patients with hypertrophy, troponin was higher: 22±21 ng/L versus 10±7 ng/L. Plasma Nt-proBNP was the same between the 2 groups 104±170 ng/L versus 64±122 ng/L. 5 patients had plasma Nt-proBNP ≥300 ng/L, they all had had atrial fibrillation or heart failure.

Plasma High sensitive troponin is a diagnostic marker of hypertrophic cardiomyopathy in Friedreich ataxia's patients, whereas plasma Nt-proBNP is associated with cardiac events and could be a prognostic marker in these patients.

The author hereby declares no conflict of interest